plastic teeth, and of these half could possibly be associated with timing of treatment.

One permanent tooth has been extracted from a patient in the study and ground sections show prominent incremental lines in dentine and abnormal morphology of the root apex with early cessation of dentinogenesis (Figure). The timing of these abnormalities are in keeping with them having occurred during the treatment for the malignant disease.

There was no difference in the hypodontia and hypoplasia between the children who had had leukaemia or solid tumours.

Discussion

The effects of chemotherapeutic agents on the developing dentition have been extensively studied in animals, but only once in humans.⁴ No abnormalities were found in the soft tissues in the present study, and the degree of caries was no more than in the general population. No abnormality of occlusion or mouth opening was seen, although this might have been expected in children with leukaemia whose temporomandibular joint would have been close to the cranial radiotherapy field. The increased incidence of hypodontia and hypoplasia can only be explained in part by the effects of chemotherapy or the disease itself. Many of the abnormalities must have occurred at times outside of the treatment period. In animals there is a variable response of the

teeth to chemotherapy,⁵ and it is likely that a similar variability would occur in man, so hypodontia and hypoplasia are not inevitable consequences of cancer chemotherapy. Indeed we have seen many completely normal teeth which must have developed during the period of cancer chemotherapy. The increased incidence of hypodontia may reflect delayed rather than absent tooth development, and only further follow up will elucidate this. In spite of these abnormalities and in view of the severe acute oral complications from which many children suffer during chemotherapy we were agreeably surprised at how healthy these children's teeth were.

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Correspondence to Dr A W Craft, Children's Department, Royal Victoria Infirmary, Newcastle upon Tyne, NE1 4LP.

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Placental steroid sulphatase deficiency

A A ATTENBURROW, M HESLIP, M J HENDERSON, J B HOLTON, I V SCOTT, AND L J H ARTHUR

Departments of Paediatrics and Obstetrics, Derby City Hospital and Department of Clinical Chemistry, Southmead General Hospital, Bristol

SUMMARY Low maternal plasma and urinary oestrogen concentrations in pregnancy are usually indicative of fetal problems, either placental insufficiency or fetal adrenal hypoplasia. Paediatricians, however, should follow obstetricians in becoming increasingly aware that deficiency of placental steroid sulphatase activity, a condition related to X linked ichthyosis, may produce the same abnormalities.

Obstetricians first became aware of placental steroid sulphatase deficiency in 1969.¹ It is associated with low concentrations of maternal plasma and urinary oestrogens, raised values of steroid sulphates, but normal human placental lactogen concentrations, and results from the inability of the placenta to metabolise steroid sulphates. The differential diagnosis is placental insufficiency or fetal adrenal hypoplasia, both conditions with a worrying prognosis. There is an association with delayed onset of labour, poor progress during labour, and failure to

breast feed.² There is a strong association with X linked ichthyosis as either the two genes are closely linked on the X chromosome or the enzyme defect causes the skin lesions.³⁻⁵ The gene seems to be on the distal tip of the short arm of the X chromosome, which includes the Xg blood group gene. These genes escape X inactivation. The inheritance is X linked recessive.

We describe two families with this condition, detected on the basis of low urinary or plasma oestriol concentrations, or both, during pregnancy.

Case reports

Family A. The mother (age 25 years, para 2+0) presented with an antepartum haemorrhage at 37 weeks' gestation. She had low serum oestriol concentrations on two occasions (less than 50 nmol/l (14 µg/l), normal more than 250 µmol/l (72 µg/l)) and a low urinary oestriol concentration (14 µmol/24 hrs (4 mg/24 hrs), normal 60 to 250 µmol/24 hrs (17 to 72 mg/24 hrs)). Human placental lactogen and urinary pregnanediol values were normal and the lecithin sphingomyelin ratio of the amniotic fluid was satisfactory. An elective caesarean section was performed as a result of short stature and a previous section. A boy was delivered weighing 3.43 kg and by the age of 3 months he had developed ichthyosis. He was bottle fed. The cord cortisol concentration was normal, excluding adrenal hypoplasia. The placenta and a fibroblast culture of the child's skin showed absence of steroid sulphatase activity when assayed by the method of Shapiro.⁶

An elder sibling had been born at term by caesarean section after failed induction of labour. His birthweight was 3.04 kg and he had ichthyosis. Skin fibroblast culture of this child and a maternal uncle with ichthyosis showed steroid sulphatase deficiency. The family tree is shown in the Figure.

Family B. The mother (age 21 years, para 2+0) was uncertain of her dates as the child was conceived shortly after stopping the contraceptive pill. There had been poor weight gain at 37 weeks' gestation (by early scan). Low plasma oestriol concentrations (54 and 70 nmol/l (15 and 20 µg/l), normal more than 250 µmol/l (72 µg/l)) had been found on two occasions but the human placental lactogen value was normal. Labour was induced at 38 weeks' gestation by syntocinon and amniotomy after 'ripening' of the unfavourable cervix with prostaglandin vaginally. A boy weighing 3.63 kg was born after a 16 hour labour. He was breast fed. The parents refused further investigation of the child, who, when seen at the age of 1 year, had developed ichthyosis. The placenta

Family A

Family B

Figure Family trees showing the inheritance of X linked ichthyosis and proved steroid sulphatase deficiency in these families.

showed reduced steroid sulphatase activity (assayed by Dr R E Oakey).

This child's elder sibling had been born at 40 weeks' gestation by forceps delivery and weighed 3.5 kg. He was breast fed and has also developed ichthyosis. This family tree is also shown in the Figure.

Comment

We describe these cases so that paediatricians may be aware of the association of placental sulphatase deficiency and X linked ichthyosis, and that undue concern is not generated by the finding of low oestrogen concentrations during such pregnancies. Parents may be reassured by a full explanation of the features of the enzyme deficiency, so that they have fewer anxieties during future pregnancies, are aware of the probability that the child may develop the associated skin changes, and know to seek appropriate advice.

We thank Dr R E Oakey, Department of Chemical Pathology, Leeds for his assistance in performing the assays on the second family.

Dr L J H Arthur died in December 1983.

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Correspondence to Dr A A Attenburrow, Green Wood Children's Centre, Nottingham City Hospital, Hucknall Road, Nottingham.

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Birthweights of infants born before 30 weeks' gestation

O G BROOKE AND N McINTOSH

Neonatal Unit, St George's Hospital, London

SUMMARY We give birthweight data with centiles on 222 singleton infants born between 24 and 30 weeks' gestation at St George's Hospital, London.

Few data are available on the weights of liveborn infants of less than 30 weeks' gestation. It is, therefore, difficult to assess whether these very immature infants are unexpectedly large or small. We provide some data which may be of use to paediatricians caring for these infants.

Patients and methods

We present data on the birthweights of all singleton liveborn infants of 30 weeks' gestation or less born at this hospital and admitted to the Neonatal Unit between February 1980 and February 1984. All infants were weighed naked and without attachments on a Marsden spring balance (Marsden Weighing Machines, London). Measurements were made to an accuracy of plus or minus 5 g. Gestation was assessed by maternal dates and antenatal ultrasound when available.

Results and comment

The Table gives the birthweight (mean (SD)) for each week of gestation between 24 and 30 weeks. There were small sex differences which did not reach statistical significance, so we have combined the data for both sexes. The Figure is a centile chart covering the same period, again with both sexes

Table Birthweights of liveborn infants between 24 and 30 weeks' gestation (both sexes combined)

	Gestation (weeks)						
	24	25	26	27	28	29	30
Number	17	12	37	32	44	33	47
Mean birthweight (g)	634	845	891	1008	1062	1212	1394
Standard deviation	81	98	151	167	186	239	327

combined. We were unable to show the bimodal distribution of birthweight or the widening of the 75th and 90th centiles found by Milner and Richards¹ in infants of less than 33 weeks' gestation. The conclusion drawn by these authors was that the higher weight peak of the bimodal distribution curve was largely formed by pregnancies of inaccurately timed gestation. It seems likely that this was so, since the absence of this phenomenon in our sample may be explained by improvements in the ultrasound assessment of pregnancies of uncertain length in the intervening 10 years. Another piece of supporting evidence is the difference in mean birthweight at 28 weeks' gestation between our sample and Milner's—his unselected infants being about 260 g heavier at that apparent gestation. It is unlikely that demographic differences would account for such a large effect, and we prefer to ascribe it to a reduction in the number of wrongly dated and more mature infants. Our mean birthweight and its standard deviation at 28 weeks' gestation was, however, similar to Babson's results from Oregon.²